Lightening the Load:

Easing the burden of constipation in children with Hirschsprung's Disease andAnorectal Malformations

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Surgical Indications and Continuity for Kids (SICK) ECHO 2018-2019
Lights on Up: Easing the Burden of Constipation in Kids with Anorectal Malformations
4/18/2019
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Provided by the University of Wisconsin-Madison Interprofessional Continuing Education Partnership (ICEP)

Intended Audience:
Primary care physicians

Objectives:
As a result of this educational regularly scheduled series, learners will be able to:
1. Assess and correct physiological and psychological problems that may increase surgical risk for regional pediatric patients.
2. Give the patient and significant others complete learning and teaching guidelines regarding the surgery.
3. Instruct and demonstrate exercises that will benefit the pediatric patient postoperatively.
4. Plan for discharge and any projected changes in lifestyle due to the surgery.

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Review of ARM (Anorectal Malformations)

Hirschsprung's Disease

- 1 in 5000 births
- Typically identified as neonate
- Ideally have surgery in first week of life

Imperforate Anus

- 1 in 5000 births
- Typically identified as neonate
- Ideally have surgery in first week of life

Cloaca – fusion of rectum, vagina and urinary tract

- 1 in 5000 births
- Typically identified as neonate
- Ideally have surgery in first week of life

Anal stenosis

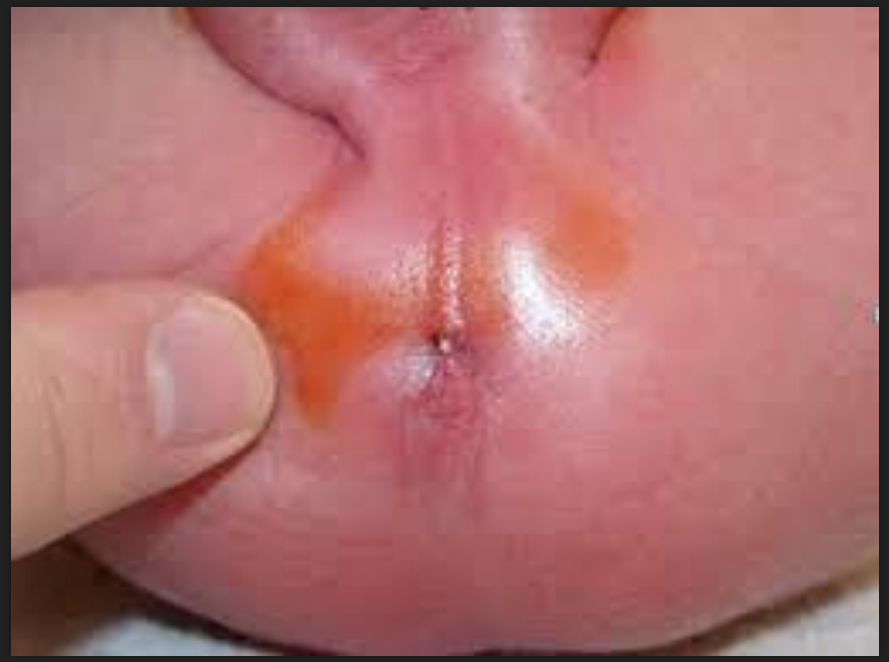
- Less than 1% of all ARM's
- Narrowing of anal canal
- Typically treat with anal dilations

A bit about me…

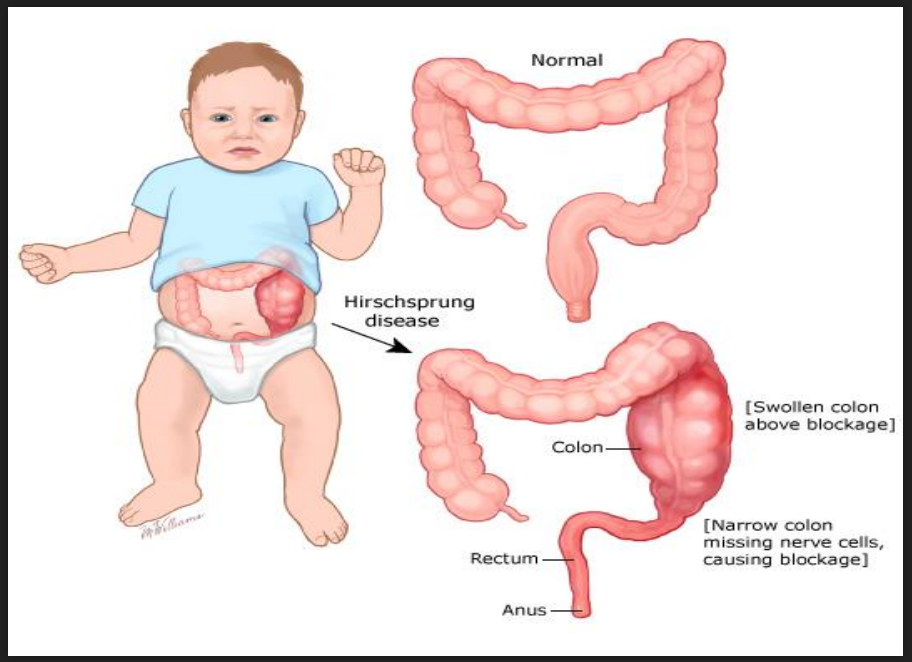
- 17 years experience in pediatrics
- 13 years as an NP – primary care, hospitalist, ED and now pediatric surgery
- I have no disclosures or financial agreements
- We are still paying off student loans
- I still owe thousands of dollars on a used Honda

Congenital obstructive defects

Imperforate Anus – a spectrum of defects

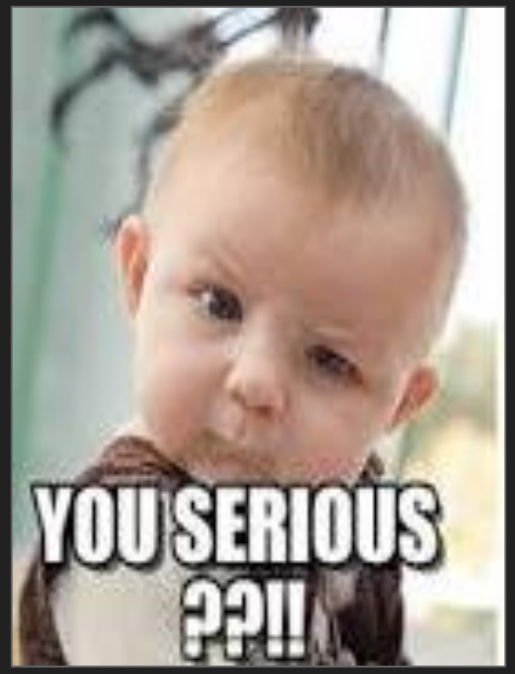


Hirschsprung's Disease – a spectrum of how much colon is affected



Typical post-op course and follow up for kids with HD/ARM's

- If defect identified as a neonate and have surgical repair within a week of age, usually < 2 week stay
- If defect is identified as an older baby, may still have a relatively short stay
- If defect is identified as a toddler/preschool/school age child, often a much longer stay and poorer prognosis
- All patient go home eating a regular diet for age (avoiding constipating foods) and getting OTC meds for pain, if any
- Nothing per rectum until f/u with surgeon!!! Zilch!
- 2 weeks post-op, begin anal dilations – most important part of post op care! Anal dilations start twice daily and taper over 3-6 months
- REGULAR FOLLOWUP: Every 2-3 months until 1 year of age, then every 3-6 months until potty trained, then every 6-12 months until middle school aged



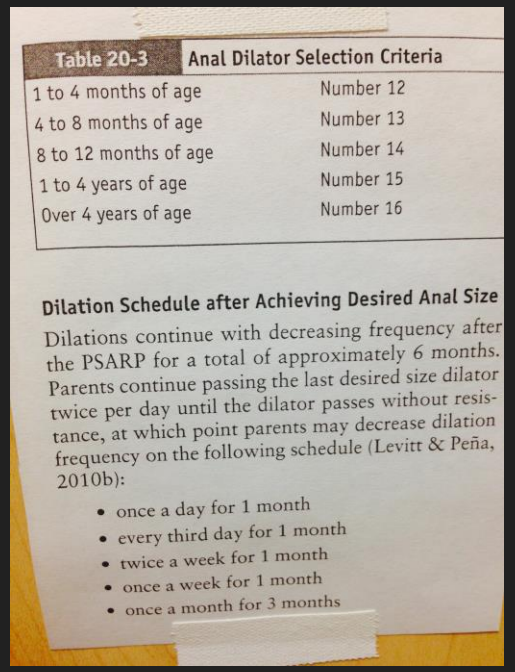


Table 20-3 Anal Dilator Selection Criteria

1 to 4 months of age	Number 12
4 to 8 months of age	Number 13
8 to 12 months of age	Number 14
1 to 4 years of age	Number 15
Over 4 years of age	Number 16

Dilation Schedule after Achieving Desired Anal Size

Dilations continue with decreasing frequency after the PSARP for a total of approximately 6 months. Parents continue passing the last desired size dilator twice per day until the dilator passes without resistance, at which point parents may decrease dilation frequency on the following schedule (Levitt & Peña, 2010b):

- once a day for 1 month
- every third day for 1 month
- twice a week for 1 month
- once a week for 1 month
- once a month for 3 months

Ongoing bowel management for HD/ARM

Pediatrician's role

- Regular well checks and all vaccines at usual intervals(including rotavirus, please!)
- Super close eye on weight trend
- Extra guidance on age-appropriate feeding with avoidance of grains/bananas/apples when starting solid foods,
- Notify Pediatric Surgery if trouble with weight gain, constipation, or diarrheal illness
- Typical management of mild constipation will not work in HD/ARM so please call us!

Pediatric surgeon/NP's role

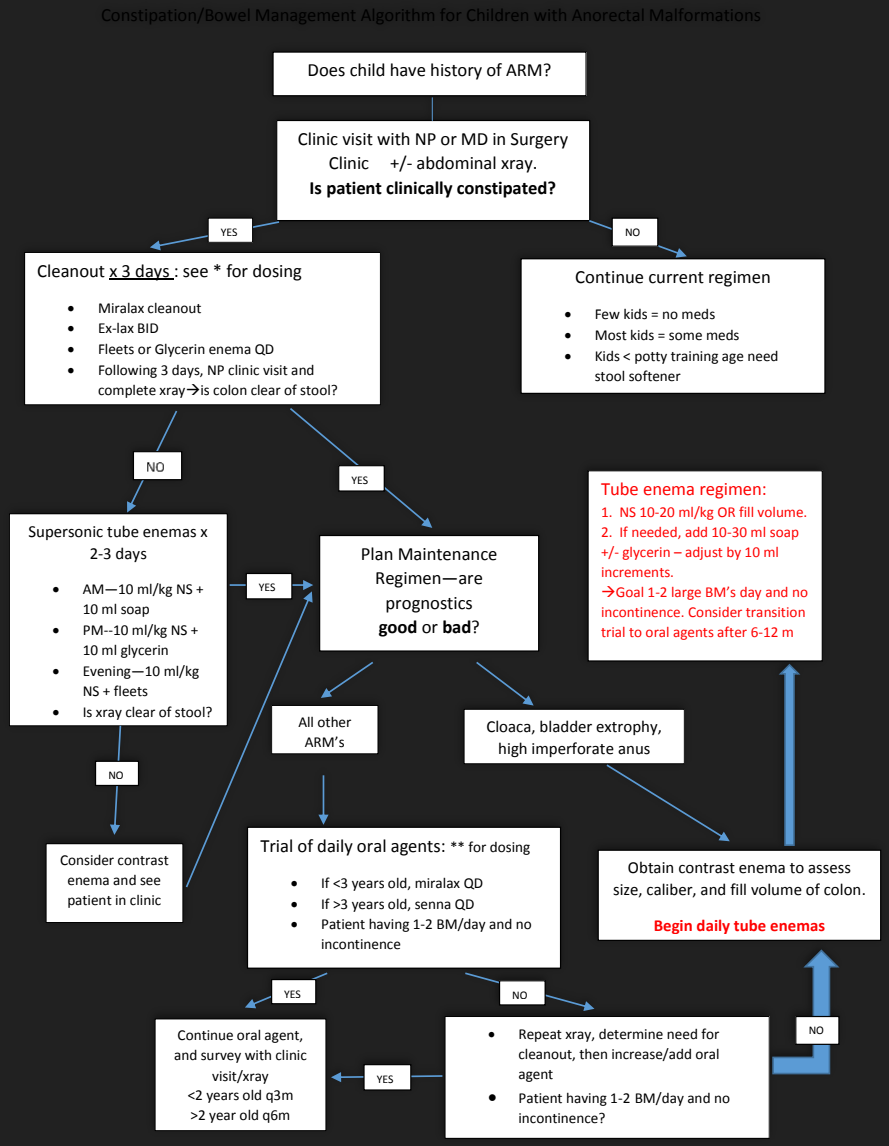
- Regular post-op follow up at specific intervals (usually every 3 months)
- Extra eye on weight gain
- Encourage starting solid foods at usual age with avoidance of grains/bananas/apples
- Manage any constipation, prescribe laxatives/stool softeners, and escalate treatment if needed
- Manage (over phone or in hospital) any diarrheal illnesses such as HAEC

What should a normal BM pattern be for a child with HD/ARM?

- Breastfeeding infants – 1 to 8 stools per day, but at least daily
- Formula fed infants – 1 to 3 stools per day, and always daily
- Toddlers /Preschoolers/School aged children – 1 to 3 stools per day, and always daily
- Kids with HD/ARM are NOT allowed to skip a day or two without stooling
- Kids with HD cannot have a vomiting/GE illness without considering possibility of HAEC
- In our experience, kids with IA tend to have more trouble than HD

Bowel management in Pediatric Surgery: how does it work?

- Regular surveillance visits with pediatric surgery every 3 months
- Usually no medications/stool softeners until toddlers unless having trouble in infancy
- Start with Miralax (PEG) ¼ to ½ cap per day in babies
- Once reaches potty training age, try withdrawing the stool softener
- If develops constipation before 1 year old or after withdrawing PEG, then use adjunctive medications



Can I get an AMEN?

Goals of Bowel Management for Pediatric Surgery

- Prevent recalcitrant constipation in children with ARM's and to achieve typical continence pattern for children who are potty training age and older – 1 to 3 BM's a day with no incontinence
- Reduce remote management of constipation in children with ARM's, while maximizing quality of care, and minimizing excessive burden of phone calls from families on RN's and NP's
- Use an algorithm that can be used to standardize guidance

Principles of Bowel Management for Pediatric Surgery

- It is prudent to stool-soften in infants and toddlers to reduce chance of developing longstanding constipation and colonic redundancy – then trial off stool softeners after potty training
- Proactively see all ARM children every 3 months for surveillance visits (with surgeon or NP) until 2 years of age (not manage remotely)
- Provide guidance/management to achieve continence at potty training age – typically around 2-3 years of age, and see them every 6 months for surveillance visits at minimum, more frequently if they are difficult to manage

Therapies used in bowel management

Medications

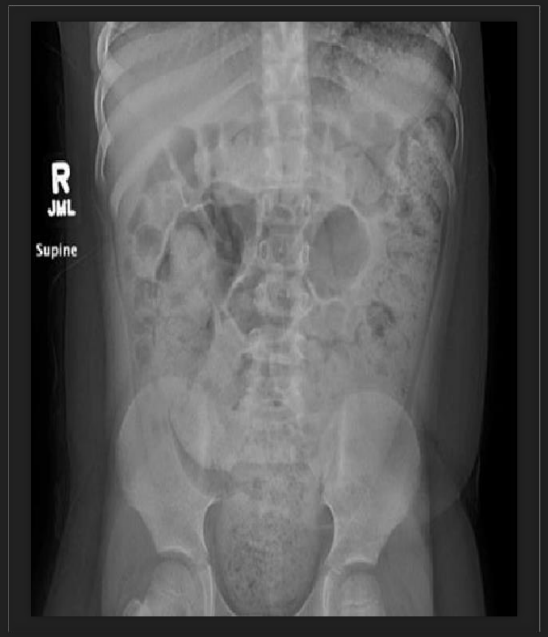
- --Miralax
  - 6 months to 1 year = ½ cap per day, titrate up if needed
  - 1-5 years = 1 cap per day, titrate up if needed
  - 6 years and up = 2 caps per day
- --Ex-lax/Senna
  - 6 months to 2 years = not typically used
  - 2-4 years = 1 tsp/day or ½ chocolate square daily, titrate up if needed
  - 5 years and up = 1-4 squares daily, even more as needed

Everything else

- OTC glycerin liquid suppositories
- Fleets or Saline enemas (child or adult size)
- Toilet hygiene
- Tube enemas
- Surgical management – MACE, cecostomy, colonic resection, or ostomy creation (last resort)

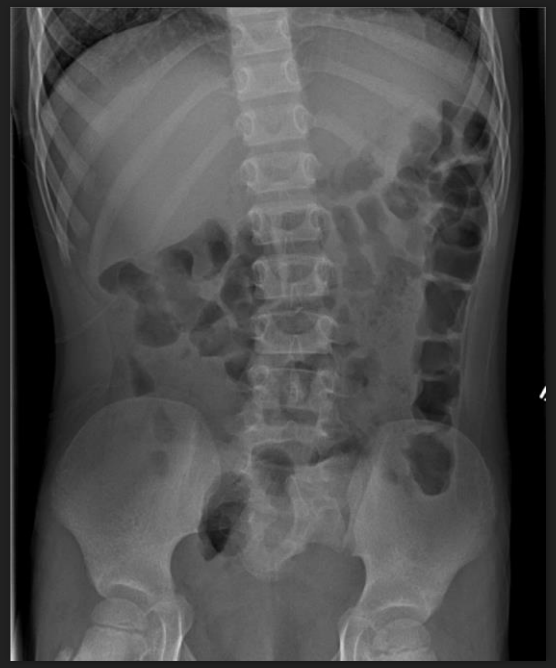
Did you poop today?

You're full of it…time for a cleanout



- 1-2 years = 3 caps Miralax in 24 oz of clear liquid,, 1 baby fleet or glycerin enema daily x 2-3 days
- 3-4 years = 5-6 caps Miralax in 40 oz of clear liquid, 1 ex-lax square BID, 1 child size fleet enema daily x 2-3 days
- 5 years and up = Miralax, 10 caps in 64 oz of clear liquid, 1-2 ex lax squares BID, 1 child or adult size fleet enema daily x 2-3 days

Show me the proof…then restart regimen



If you fail, get a Barium Enema and try again!

This is NOT normal – time to initiate enema therapy

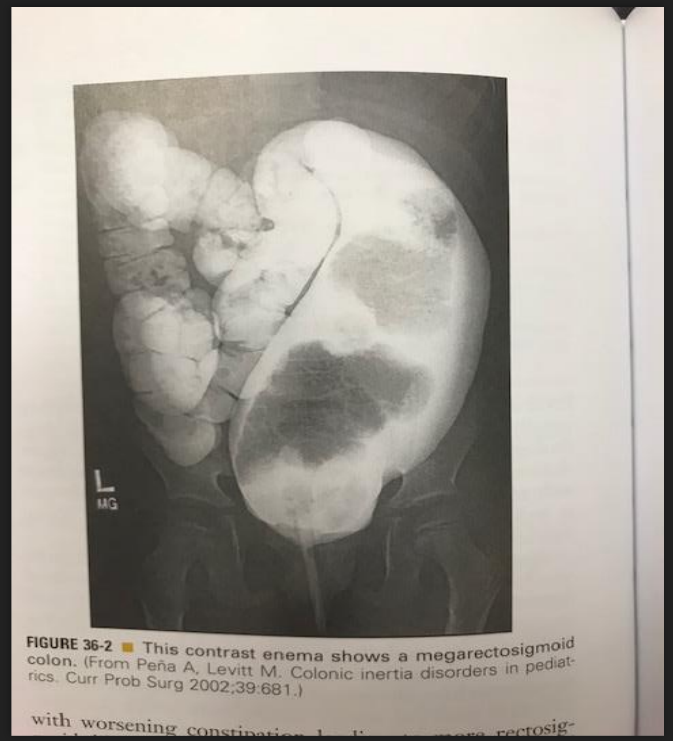


FIGURE 36-2 ■ This contrast enema shows a megarectosigmoid colon. (From Peña A, Levitt M. Colonic inertia disorders in pediatrics. Curr Prob Surg 2002;39:681.)

with worsening constipation, [illegible] rectosig-

After 6 month trial of large volume tube enemas

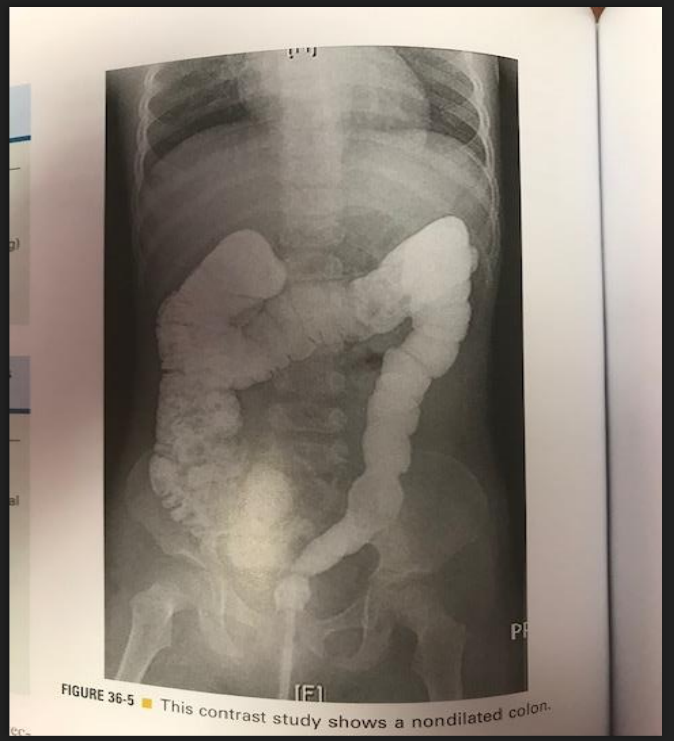


FIGURE 36-5 ■ This contrast study shows a nondilated colon.

Tube enemas for bowel management

- Not the same as OTC small volume enemas
- Fill the colon with enough solution to liquefy any solid stool – use "fill volume" from BE
- Start with saline and add glycerin or soap only if needed
- No limit on volume for enemas
- Once daily for 6 months, then trial off if verify clean colon on xray
- When withdrawing enemas, will need to start senna dosing QD

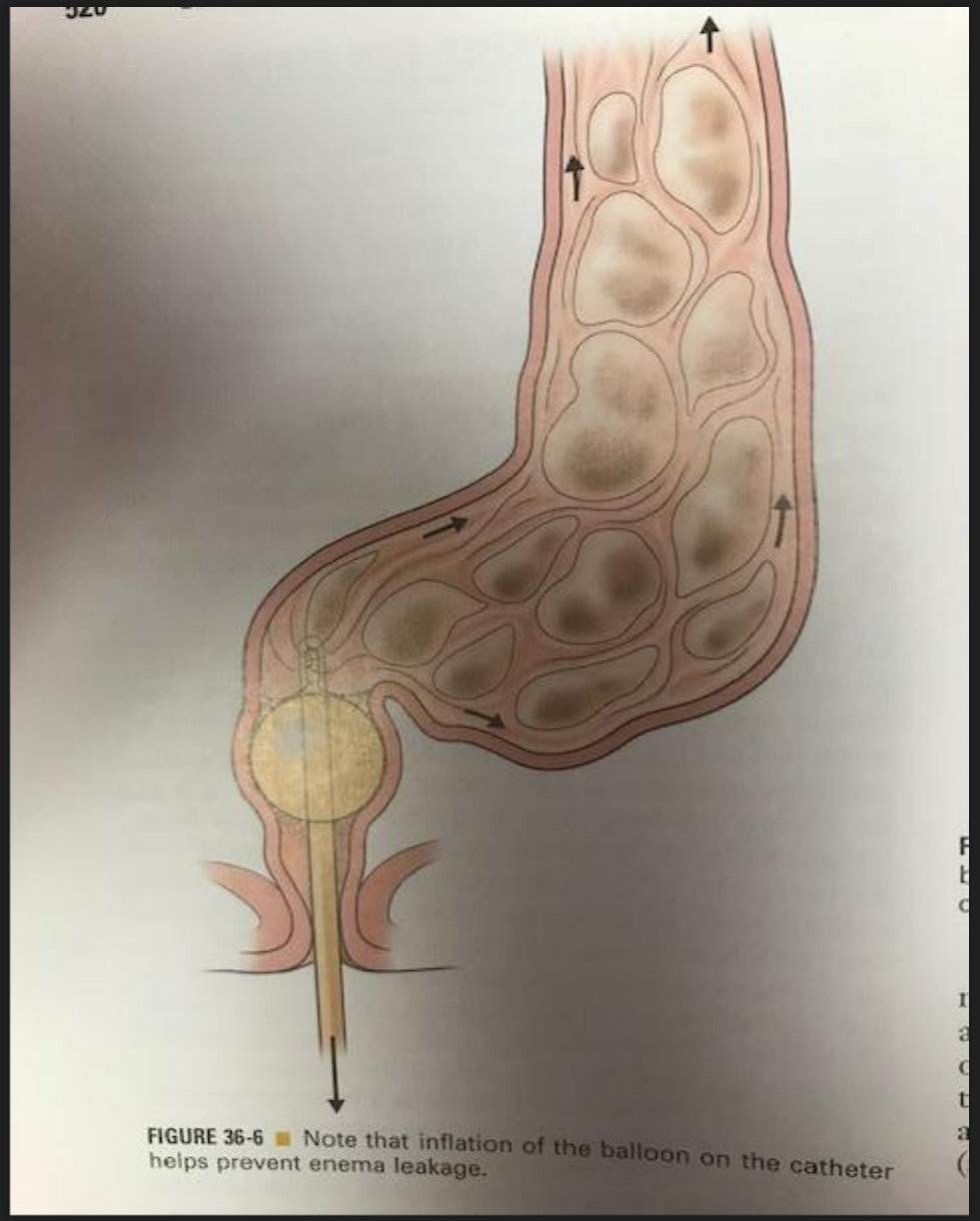
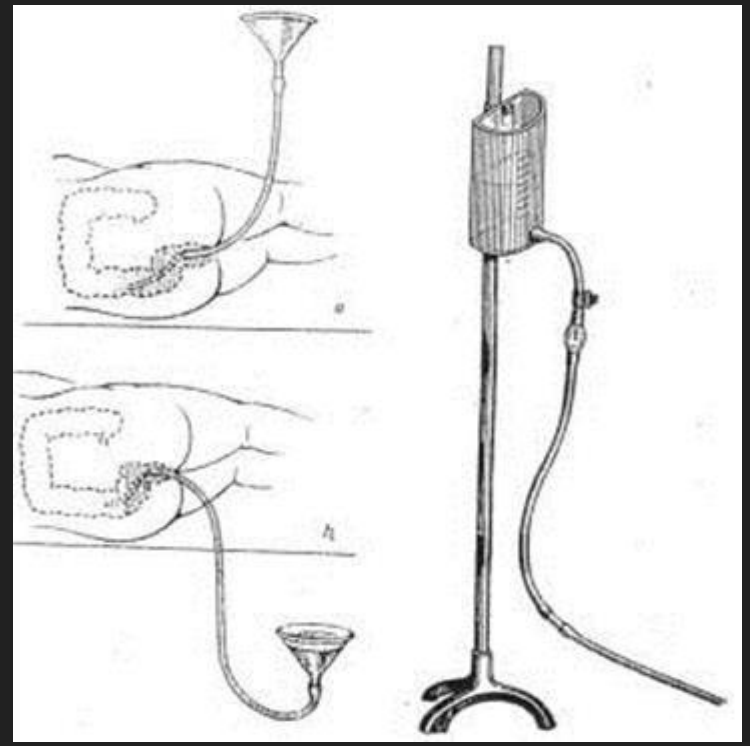


FIGURE 36-6 ■ Note that inflation of the balloon on the catheter helps prevent enema leakage.

What about Peristeen?

- Device for daily, long term enemas
- Same premise as tube enemas but comes in a kit
- Rarely covered by insurance, but getting better – cost is $1000-2000 per year
- Rectal tube and gravity bag = $2 per year
- We have no patients who use this but you are welcome to try



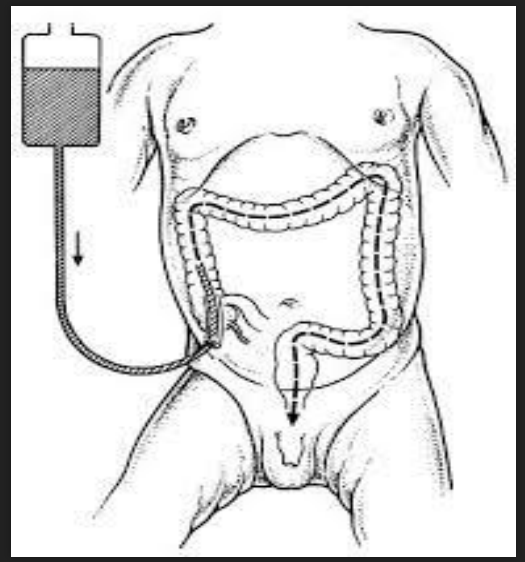
Referring to a specialty center: When does the surgeon phone a friend?

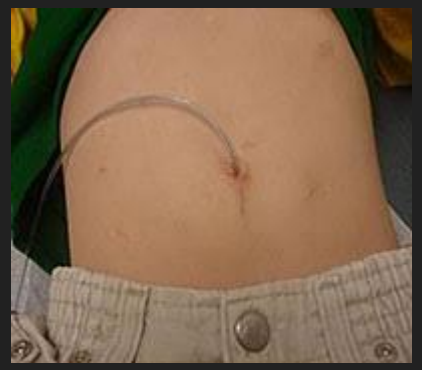
Nationwide Children's Hospital or Children's Hospital of Colorado, Cincinatti Children's Hospital

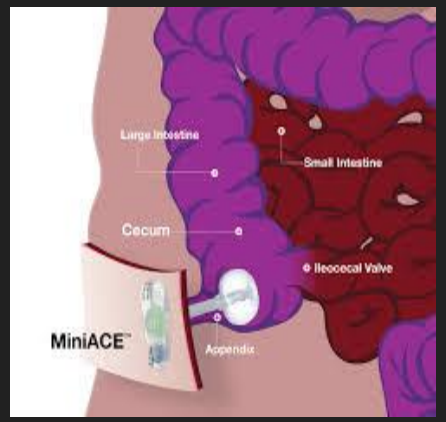
- When bowel management algorithm has been exhausted
- Failed multiple adjustments of medications
- Tube enemas are not working
- Failure to wean off tube enemas at school age
- Need colonic manometry or small bowel motility studies
- Considering bowel resection of ostomy
- May need repeat PSARP or pull through
- Cloaca/complex defects

Escalating bowel management: MACE, ostomy, bowel resection – OH MY!

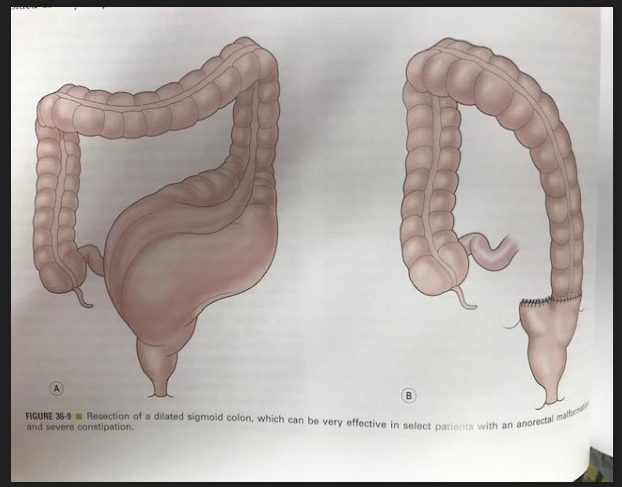
MACE or Cecostomy







Sigmoid colon resection



Bowel management for HD/ARM: Take home messages for primary care providers

- Inquire about BM pattern at every primary care visit
  - > 1 year of age: 1 to 3 BM's per day
- Make sure patients are getting age appropriate follow up with pediatric surgery
  - Babies/toddler q 3 months
  - Preschool/school age q 6 months
- Recognize that bowel management is a HUGE BURDEN and that early intervention is key
- Remember the red flags
  - Frequent smearing, accidents, skid marks, decreased weight acceleration, frequent stooling